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Titanium plating system with autologous rib graft sternoplasty in the treatment of thoracic inlet compression

Matthew Alexander [a], John Keech [b], Peter J. Gruber [b], Donald Dean Potter [c,*]

[a] Department of Surgery, University of Iowa Hospitals and Clinics, Iowa City, IA, USA
[b] Department of Cardiothoracic Surgery, University of Iowa Carver College of Medicine, Iowa City, IA, USA
[c] Division of Pediatric Surgery, Mayo Clinic, 200 First Street SW, Rochester 55905, MN, USA

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ABSTRACT

Narrowing of the thoracic inlet leading to airway compression is a rare and challenging condition in the pediatric population. Reports in the literature have described this variant related to multiple conditions including double crush phenomenon following repair of pectus excavatum, anterior spinal displacement, and straight back syndrome. Underlying genetic conditions such as Marfan's Syndrome and Hurler's Syndrome have also been reported to contribute to clinically significant airway compression independent of dynamic tracheal collapse such as tracheomalacia. The borders of the thoracic inlet are anatomically bound by the body of the first thoracic vertebrae (T1) posteriorly, the posterior surface of the manubrium anteriorly, and the medial aspects of the first ribs on either side laterally. Relief of tracheal compression in this location is complicated by the rigidity of the boney thoracic inlet and limited space for lifting procedures such as anterior aortopexy. Several operative approaches to treat this condition have been described including manubrial/sternal resection, first rib resection, and reconstruction of the thoracic inlet. Described here are three patients where successful reconstruction of the thoracic inlet was achieved using autologous rib graft sternoplasty and a titanium sternal plating system to widen the thoracic inlet and eliminate external compression on the trachea.

Narrowing of the thoracic inlet leading to airway compression is a rare and challenging condition in the pediatric population [1]. This unusual phenomenon has been associated with conditions including anterior spinal displacement, straight back syndrome, and double crush phenomenon [2–4] as well as various genetic diseases [5–8]. Due to the complexity of the anatomy, several surgical approaches have been described including sternal resection, first rib resection, and reconstruction of the thoracic inlet [7,9–12]. Here we describe three patients in whom tracheal compression was successfully relieved by utilizing autologous rib graft and titanium sternal plating to expand the anterior boundaries of the thoracic inlet and relieve tracheal compression.

1. Case reports

Patient 1 was a 14-year-old boy with a history of hypoxic ischemic encephalopathy and spastic cerebral palsy. Six months following a laparoscopic fundoplication with gastrostomy tube for gastroesophageal reflux disease, his family described worsening cough, gagging, and choking. He was evaluated by otolaryngology and underwent a tonsillectomy and adenoidectomy for upper airway obstruction. Two weeks following this procedure, he experienced a hypoxic arrest at home. He was intubated and transferred to our institution. He failed extubation twice due to repeated acute life threatening events secondary to airway obstruction. Diagnostic bronchoscopy revealed non-dynamic external compression of the distal trachea. The left mainstem bronchus was not able to be intubated due to external compression. Chest computed tomography showed there was significant compression of the trachea and left mainstem bronchus by the innominate artery and aortic arch (Fig. 1A). On sagittal images, the distance of the thoracic inlet was 28 mm, likely due to a loss of

* Corresponding author. Tel.: +1 507 284 8391; fax: +1 507 284 0058.
E-mail address: potter.donald14@mayo.edu (D.D. Potter).

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Fig. 1. (A) 3-D reconstruction of chest computed tomography demonstrating severe trachael compression just distal to the endotracheal tube and of the left mainstem bronchus. (B) Pre-operative chest computed tomography showing the loss of normal thoracocervical kyphosis consistent with straight back syndrome and trachael compression by the aortic arch. The distance from the posterior manubrial plate to the anterior vertebral line is 28 mm.

normal thoracic kyphosis consistent with straight back syndrome (Fig. 1B). Consequently, a standard aortopexy was unlikely to relieve the tracheal compression.

The patient was taken to the operating room for median sternotomy, transposition of the innominate artery and aortopexy with intra-operative bronchoscopy [4]. Once the sternum was opened, the compression on the trachea was relieved as determined by intraoperative bronchoscopy. The relief was augmented by lifting the innominate artery and aorta anteriorly. However, the compression recurred when the sternal edges were reapproximated. Thus, the decision was made to perform a rib expanded sternoplasty instead of transposing the innominate artery. The 6th rib was harvested and divided into three 2 cm sections. The upper sternum was expanded using the 2 cm rib struts and stabilized with titanium plates (Fig. 2).

The aorta was secured to the posterior sternal plate with permanent thread prior to closure. The lower sternum was closed in standard fashion with wire. Pectoralis major advancement flaps were mobilized bilaterally to cover the defect. The tracheal compression was markedly improved upon completion of the reconstruction. He was able to be extubated on postoperative day 3 without recurrent airway symptoms. His hospital stay was extended due to post-cardiotomy syndrome treated with steroids. On follow-up 6 months later, he remains symptom free from airway obstruction. Repeat chest computed tomography demonstrates expansion of the thoracic inlet from 28 mm preoperative to 35 mm postoperatively (Fig. 3). Due to concern for worsening thoracic lordosis, an orthopedic consultation was obtained and continued observation was recommended.

Patient 2 was a 7-year-old girl with 15q partial trisomy and 9p partial monosomy syndrome with developmental delay and hypotonia. She had been followed for 18 months by the pulmonary allergy group for persistent bacterial bronchitis. Chronic antibiotics, reflux medications, and anti-inflammatory medications failed to relieve her symptoms and her cough worsened to such an extent that she was removed from her classroom. On exam, she had marked manubriosternal synostosis (pouter pigeon breast) with a manubriosternal angle of 135° by chest x-ray. Chest computed tomography displayed normal vasculature and confirmed the sternal deformity with tracheal compression (Fig. 4A). Diagnostic bronchoscopy found severe compression at the carina involving the left mainstem bronchus with an estimated 80% loss of circumference. Airway pressures of 35–40 mm Hg resulted in expansion to 50% of the normal circumference. This finding along with the CT scan suggested that aortopexy alone would not relieve the tracheal compression.

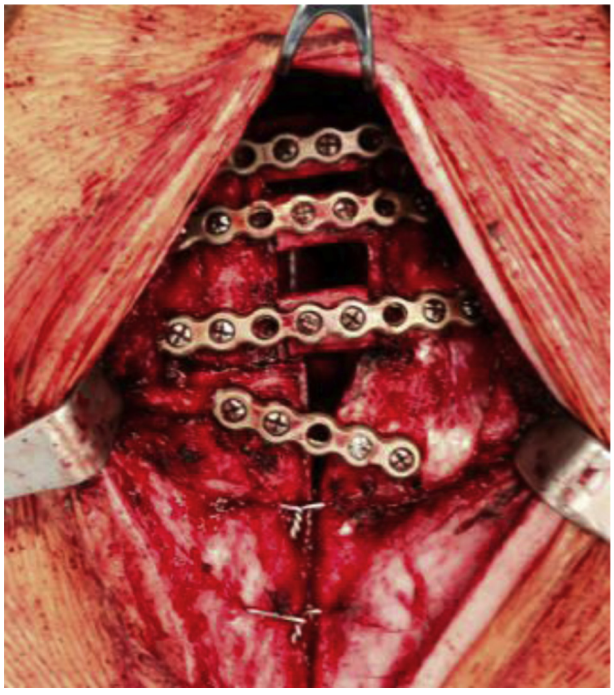

Fig. 2. Intra-operative photography of the titanium plated rib expanded sternoplasty. The patients head is cephalad. The 3–2 cm rib segments are secured with titanium plates to each side of the sternum. The lower sternum is reapproximated using wire.

Fig. 3. Postoperative chest computed tomography scan demonstrating the increased thoracic inlet. The distance from the posterior manubrial plate to the anterior vertebral line is now 35 mm.

The patient was taken to the operating room for a modified Ravitch procedure with intra-operative bronchoscopy. Upon elevating the sternum, the tracheal compression was relieved. However, we were not able to secure the manubrium in the elevated position and were not willing to place a posterior manubrial strut at this level due to the proximity of the great vessels. Consequently, we proceeded with a partial sternal split and a rib expanded sternoplasty stabilized with titanium plates. The aorta was secured to the posterior sternal plate with permanent thread and the manubrium was elevated with vertical titanium plates (Fig. 4B). Bilateral pectoralis advancement flaps covered the defect. This provided a stable repair and relieved the compression. She was extubated on postoperative day 2 and has had an uncomplicated course. At 6 months follow-up, she no longer has a cough, has returned to school, and has had no further infections. Chest x-ray shows a normal contour of her sternum with increased thoracic inlet diameter (Fig. 4B).

Patient 3 was 17-year-old male with Marfan's syndrome. At age 13 he underwent posterior fixation for severe kyphoscoliosis. Postoperatively he developed severe exercise intolerance with marked hypercapnea during exercise testing. He underwent extensive cardiorespiratory testing but this failed to reveal an explaination for his dyspnea on exertion. He eventually underwent chest computed tomography to evaluate the mechanism. Severe pectus excavatum was identified in addition to tracheal compression at the level of the innominate artery, and aortic arch; additionally, he had aortic root dilation. While the diagnosis of double crush syndrome was considered [2], restricted thoracic inlet causing tracheal compression due to acquired straight back syndrome was felt to contribute significantly to his symptoms.

Aortic root replacement was indicated due to his aortic aneurysm with insufficiency. He underwent aortic root replacement with a 25 mm St Jude valved conduit. In addition, innominate arterial compression was relieved by extension of the artery using a Goretex interposition graft between the native innominate artery and aorta. The airway was examined intra-operatively and revealed distal tracheal compression when the chest wall was closed. Thus, rib expanded sternoplasty stabilized with titanium plates was utilized as the prior two patients with near resolution of tracheal compression. His postoperative course was uncomplicated. On postoperative follow up, his breathing has improved but he continues to have some dyspnea on exertion. Repeat chest computed tomography illustrates an increase in the thoracic inlet size from 28.0 mm to 42.5 mm (Fig. 5).

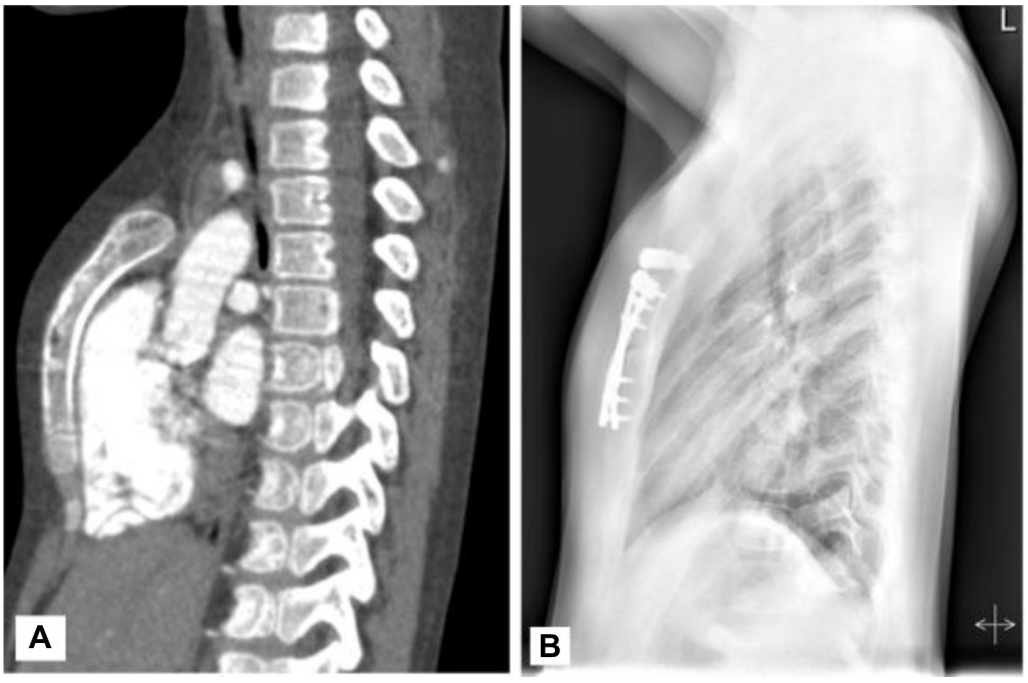

Fig. 4. (A) Pre-operative chest computed tomography demonstrating the manubriosternal synostosis (pouter pigeon breast) and tracheal compression. (B) Post-operative chest roentgenogram depicting the increased thoracic inlet following rib expanded sternoplasty with titanium plating.

2. Discussion

Thoracic inlet narrowing can cause significant tracheal compression and is an unusual diagnostic and therapeutic dilemma facing the pediatric surgeon. Multiple abnormal anatomic variants have been associated with this condition in the literature. These include acquired straight back syndrome, pectus excavatum with double crush phenomenon, and anterior spinal displacement. Each of these disorders constrict the normal bony borders of the thoracic inlet, which may lead to external tracheal compression.

Anterior aortopexy is one strategy used to reduce collapse of the trachea and provide symptomatic relief by pulling the aorta away from the airway. Two common approaches to this procedure may be utilized based on the direction of compression on the trachea. Classically, aortopexy is performed by leaving the pretracheal fascia intact, which allows the tracheal wall to be pulled forward with the aorta to expand the lumen. When compressed by vascular structures, aortopexy can be achieved by dissecting the pretracheal fascia from the posterior aorta to relieve pressure by the vascular structures [9]. However, both of these maneuvers are limited by the distance between the sternum and the aorta. As seen in these three cases, extreme narrowing of the thoracic inlet excludes aortopexy as a complete treatment option. In this scenario, it is necessary to expand the bony borders of the abnormally small thoracic inlet.

The traditional approach to expanding the thoracic inlet is to remove the lateral border with a first rib resection [7]. However this technique carries with it the substantial risks associated with extensive dissection and single lung airway ventilation [7]. Additionally, expansion of the thoracic inlet in the lateral dimension may not provide sufficient relief in the setting of anterior or posterior tracheal compression. Thus, a technique to expand the thoracic inlet in the anterior/posterior plane is necessary. A reconstructive maneuver to expand the thoracic inlet anteriorly has been previously described utilizing iliac crest bone graft, internal fixation plates, and pectoral muscle flaps [7]. Our approach utilizes autologous rib graft sternoplasty and a titanium plating system to provide stable anterior expansion of the thoracic inlet. Coverage of the anterior mediastinum is achieved using rib grafts, titanium plates, and bilateral pectoralis muscle flaps. Although the rigid coverage is not contiguous, we believe that the soft tissue coverage is adequate short-term protection while utilizing sternal precautions. Our standard sternal precautions include pillow hugging with laughter, coughing, and sneezing, lifting restriction to no greater than 8 lbs, and avoidance of pushing or pulling. Additional precautions include using only legs to stand and only legs or elbows to help get out of bed. We extended the standard sternal precautions for a total of six months. The six month precaution is somewhat longer than the typical precaution period of 6–12 weeks, but was applied due to the unique nature of the operation. The rib grafts are expected to create long-term stability following the initial healing period. Our current cohort of patients will not participate in competitive athletics or other high impact sports. Strong consideration to permanent contact restrictions would be given to any patient that is able to participate in such activities. This procedure was associated with few complications that were managed without difficulty and has proved to provide a stable repair in the six months post-operative period. Moreover, all three patients undergoing this procedure experienced significant symptomatic relief. Continued follow up is required to determine whether stability of the anterior chest is achieved long-term.

Fig. 5. (A) Pre-operative chest computed tomography showing the narrow thoracic inlet and tracheal compression. (B) Postoperative chest computed tomography demonstrating the increased thoracic inlet following rib expanded sternoplasty with titanium plating.

3. Conclusion

We have described a new surgical approach to reconstruct the thoracic inlet for the treatment of thoracic inlet compression. This operative strategy decompresses the thoracic inlet and improves symptoms of tracheal compression when aortopexy alone is insufficient. In our experience, reconstruction of the thoracic inlet with rib autograft and titanium sternal plating has proved to be an effective and stable technique to treat tracheal compression without significant complication.

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